



Cancer Research Center Hotline

Description of Neoplasms in Infants Diagnosed Under One Year of Age in Hawaii, 1986-2000

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Hawaii Birth Defects Program (HBDP)

The Hawaii Birth Defects Program (HBDP) was established in August 1988 as a surveillance system and registry for birth defects and other adverse pregnancy outcomes. A birth defect is defined as any structural, functional, or biochemical abnormality in development that originates before birth and is detectable at birth or shortly thereafter. These defects may be genetically linked or caused by environmental hazards or adverse lifestyle effects, but most often are of unknown origin.¹

Birth defects are the single most common cause of infant mortality in Hawaii except for other perinatal conditions and all other causes.² On average 1,000 (5%) of all babies born each year in Hawaii are diagnosed with some recognizable adverse pregnancy outcome.³ About 12% of all admissions to pediatric hospitals are caused by problems directly associated with birth defects.⁴ As a result, birth defects and other adverse pregnancy outcomes represent a significant impact for Hawaii's hospitals and families.

The purpose of the HBDP is to be a reliable, valid, and timely information source for ascertaining the number of infants up to one year of age with specific birth defects and other adverse reproductive outcomes, as well as providing relevant and timely information and data to community agencies, hospitals, and state departments for policy, planning, and decision making purposes. The HBDP works to fulfill this objective by being a statewide, population-based surveillance system that uses active ascertainment methodology and collects information from multiple sources (approximately 33 health care facilities and vital records).⁵

With its 15 years of data and statistical analyses, the HBDP has examined elevated adverse outcomes, possible geographic and other local clusters (Village Park and Royal Kunia), and birth defect trends and changes over time. At the national level, the HBDP served as the pilot site and in Phase II studies of the Department of Navy's Gulf War Syndrome Birth Defects Study.⁶ The Program also provides information for special studies and for developing public awareness education programs about birth defects and their causes. And lastly, the HBDP acts as a resource for the planning and development of appropriate statewide and community level services and preventive strategies.

From 1988 until 2002, the HBDP was covered under Hawaii Revised Statutes (HRS), sections 321-31 and 338-2, for its statutory authority, with additional legislation (HRS 324-1 and 324-2) added

in 1990 Amendments. During the 2002 Hawaii Legislative Session, Act 252 (SB 2763, SD2, HD2, CD1 specifically relating to birth defects) was enacted by the legislature, and signed by the Governor on July 1, 2002. Funding for the Program has come through federal, state, and foundation public and private sources.

The HBDP carries out its own research, culminating in data and information dissemination to community requesters, presentations, publications, and 22 articles published in professional peer-reviewed journals.³

In 1999, the Pew Environmental Health Commission at the Johns Hopkins School of Public Health, and in 2002, the Trust for America's Health, both gave the HBDP an A grade and designated it as one of the top eight programs in the country, and one of only four nationwide to meet all rating criteria.^{7,8}

Neoplasms in Infants Diagnosed Under One Year of Age in Hawaii

There are several reasons why a birth defects registry might collect information on neoplasms. Birth defects and neoplasms may have similar etiologies. For example, it has been suggested that sex hormones and their analogues might cause the birth defects hypospadias and undescended testicle as well as testicular cancer.⁹ And neoplasms may be prenatally diagnosed and electively terminated.¹⁰ Regular cancer/tumor registries do not normally identify these pre-birth cases, while birth defects registries such as the HBDP that include fetal demises, prenatally diagnosed cases and elective terminations, include them as part of their regular case ascertainment procedures. In addition, regular cancer/tumor registries typically do not include benign tumors such as lipomas. If a birth defects registry includes benign tumors, then it would serve as a source of information for such tumors where a cancer/tumor registry would not.

The HBDP collects information on neoplasms, both benign and malignant, that are diagnosed between conception and one year after delivery. Excluding 151 cases of cystic hygroma (lymphangioma), which is strongly associated with chromosomal abnormalities and may spontaneously resolve during pregnancy,^{11,12} the HBDP identified an additional 114 cases of neoplasms diagnosed before age one year among deliveries during 1986-2000. In that time period there were 281,866 total live births in Hawaii, resulting in an infant and fetus neoplasm rate of 4.0 per 10,000 births. The most common type of neoplasm was teratoma (n=21, 18.4%), followed by lipoma (n=18, 15.8%), neuroblastoma (n=14, 12.3%), retinoblastoma (n=12, 10.5%), hepatoblastoma (n=8, 7.0%), and medulloblastoma (n=5, 4.4%). Live births accounted for 108 (94.7%) of the cases, of which 12 (11.1%) were known to have died within one year of age. The remaining cases were evenly divided among elective terminations (n=3, 2.6%) and fetal deaths (n=3, 2.6%). In 16 (14.0%) of the cases, the neoplasm was prenatally diagnosed on ultrasound.

The number of infants and fetuses with neoplasms varied widely by year, with a low of two among infants delivered in 2000 and a high of 15 in 1994. There was no clear secular trend in neoplasm rates during the 15-year period (p=0.086).

Although the neoplasm rate among cases with maternal age less than 35 years (4.2 per 10,000 births) was higher than the rate among cases with maternal age of 35 years or greater (3.0 per 10,000 births), the difference was not statistically significant (p=0.162). The rates

among males (4.1 per 10,000 births) and females (4.0 per 10,000 births) were similar. Neoplasms were substantially more common among multiple births (12.2 per 10,000 births) than among singleton births (3.9 per 10,000 births) ($p=0.008$). Of the seven cases of neoplasms among multiple births, there were two pairs of twins concordant for the neoplasm (one set of retinoblastoma and one set of medulloblastoma).

Among live births, the neoplasm rate was significantly higher with gestational age at delivery of less than 38 weeks (8.8 per 10,000 births) when compared to 38 weeks or greater (2.9 per 10,000 births) ($p<0.001$) and for birth weight of less than 2,500 grams (6.6 per 10,000 births) when compared to birth weight of 2,500 grams or greater (3.5 per 10,000 births) ($p=0.024$).

The frequency of neoplasms was similar among deliveries to residents of metropolitan Honolulu (4.1 per 10,000 births) and the rest of the state (3.9 per 10,000 births) ($p=0.391$). Although the rates varied between the counties — City and County of Honolulu (4.0 per 10,000 births), Hawaii County (4.1 per 10,000 births), Maui County (2.5 per 10,000 births), and Kauai County (2.2 per 10,000 births) — the differences were not statistically significant.

Neoplasm rates were highest with maternal race/ethnicity of Pacific Islander (Hawaiian, Samoan, and Guamanian) (4.5 per 10,000 births), followed by Filipino (4.2 per 10,000 births), and white (3.9 per 10,000 births), and lowest among Far East Asians (Japanese, Chinese, and Korean) (3.7 per 10,000 births). However, the contrasts in rates were not statistically significant.

Twenty-seven (23.7%) of the infants and fetuses had a cytogenetic analysis performed. Of these, four were found to have chromosomal abnormalities — partial trisomy 2 (retinoblastoma), trisomy 18 (teratoma), mosaic trisomy 19 (leukemia), and translocation chromosomes 4 and 11 (leukemia). Fifty-four (47.4%) of the cases had major structural birth defects. The more common major structural birth defects were hydrocephaly ($n=11$), microcephaly ($n=3$), and ventricular septal defect ($n=3$). Six (5.3%) of the cases had congenital syndromes — Russell-Silver syndrome, Klippel-Trenaunay-Weber syndrome, Beckwith-Wiedemann syndrome, Kasaback-Merritt syndrome, oro-facial-digital syndrome, and Fanconi syndrome.

Further information and data from the Hawaii Birth Defects Program can be secured by contacting the Administrator, Ruth D. Merz, at 76 North King Street, #208, Honolulu, Hawaii, 96817-5157, 808-587-4120 (phone), 808-587-4130 (fax), hbdp@crch.hawaii.edu (e-mail), or <http://www.crch.org/hbdp> (website).

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- Store all medicines, chemicals, and household products out of reach and out of sight, preferably locked up.
- A good rule to teach children is to "always ask first" before eating or drinking anything—don't touch, don't smell, don't taste.

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